Congenital Choledochal Dilatation With Emphasis on Pathophysiology of the Biliary Tract

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Of 37 patients with congenital choledochal dilatation, aged 8 days to 12 years, who had undergone excision with Roux-en-Y hepaticojejunostomy, 26 patients could be analyzed for morphologic abnormalities and pathophysiology of the biliary tract. Of the 26 patients with congenital choledochal dilatation, 25 (96.2%) had an abnormal choledochopancreaticoductal junction. Of the 12 patients with cystic-type choledochal dilatation, 10 had the C-P type of abnormal choledochopancreaticoductal junction, and of the 13 patients with fusiform-type choledochal dilatation, nine had the P-C type. The amylase levels in the choledochal cyst and the gallbladder were elevated regardless of the form of choledochal dilatation. An adenocarcinoma in a cystic choledochal dilatation was found in one child. Therefore, longstanding inflammation of the biliary tract caused by the reflux of pancreatic juice might be one of the factors in carcinogenesis in the biliary tract. This free reflux of pancreatic juice was demonstrated not only by amylase levels in the biliary tract but also by intraoperative biliary manometry. This reflux might be explained by the lack of sphincter function at the junction of the common bile and pancreatic ducts.

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ADVANCES IN ENDOSCOPIC retrograde cholangiography (ERCP) in infants have provided further elucidation of the relationship between congenital choledochal dilatation and abnormalities of the choledochopancreaticoductal junction. However, little is known about the pathophysiology of the biliary tract in patients with congenital choledochal dilatation.

This paper describes morphologic abnormalities and the pathophysiology of the biliary tracts of the 37 patients with congenital choledochal dilatation. In addition, the pathophysiology, which might lead to malignant degeneration in the biliary tract, is discussed.

Clinical Data

Thirty-seven patients with congenital choledochal dilatation were treated at the Division of Surgery, Children's Research Hospital, Kyoto Prefectural University of Medicine from 1978 to 1990. The mean age of the nine male patients was 2 years 6 months (range, 8 days to 5 years). The mean age of the 28 female patients was 3 years 10 months (range, 1 month to 12 years) (Table 1). During the follow-up period of 1 to 13 years, 36 patients have been doing well. However, one patient—a 12-year-old girl[1]—died of adenocarcinoma arising from the distal part of the cyst in the infrapancreatic region 2 years after operation.

Excision of choledochal cyst followed by Roux-en-Y hepaticojejunostomy was performed in all 37 patients. The cyst was proximally transected at the hepatic hilum, and distally resected at the level of the infrapancreatic region.

The biliary tract was examined by preoperative ERCP or operative cholangiography in all 37 patients. Union of the common bile duct and the main pancreatic duct could be clearly demonstrated in 26 patients, 16 by ERCP, 3 by operative cholangiography, and 7 by both procedures. The findings were classified into three types of abnormal choledochopancreaticoductal junction: (1) C-P type, the bile duct joins the pancreatic duct, which is the major duct (Fig. 1); (2) P-C type, the pancreatic duct joins the bile duct, which is the major duct (Fig. 2); (3) miscellaneous type, the connection between the common bile duct and the pancreatic duct is too complex to be categorized as either C-P or P-C type.

Biliary manometry could be performed in the most recently treated 11 of the 37 patients. Pressure recordings were obtained with a polyvinyl catheter with an internal diameter of 0.8 mm and an outer diameter of 1.0 mm. The probe was filled with sterile solution, and perfusion took place at a constant rate of 30 mL/hour. This appa-

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TABLE 1. *Patients With Congenital Choledochal Dilatation*

Sex	No. of Patients	Mean Age (range)
M	9	2 yr, 6 mo (8 day–5 yr)
F	28	3 yr, 10 mo (1 mo–12 yr)
Total	37	3 yr, 6 mo (8 day–12 yr)

ratus was connected to a transducer (Gould Inc., P231D, Oxnard, CA), and the pressures were recorded with a Nippon-Sanei thermal pen recorder. Zero pressure, used throughout this study, was determined by recording atmospheric pressure at the distal end of the common bile duct. To obtain pressure recordings from the biliary duct system, the probe was inserted into the sphincter of Oddi through the distal end of the common bile duct and the abnormal choledochopancreaticoductal junction. The probe was withdrawn from the duodenum to to the biliary tract at a constant speed of 0.8 mm/second. The pressure profile of the biliary tract was recorded in centimeters as the probe was withdrawn. Tetragastrin (4 r/kg) was injected intravenously, and manometric studies of the biliary tract were performed before and 3 minutes after the injection.

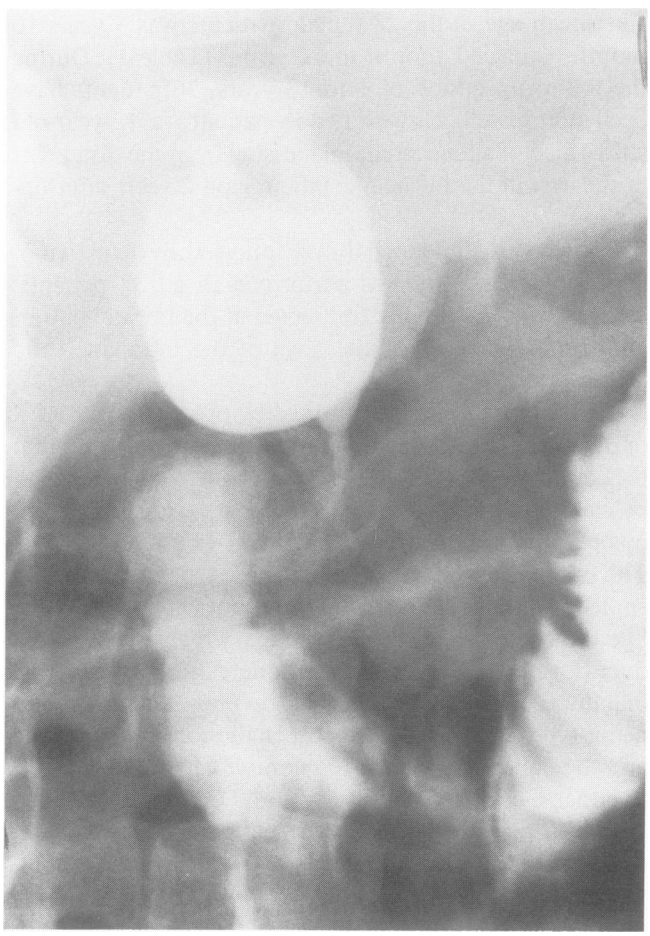

FIG. 1. C-P type of abnormal choledochopancreaticoductal junction. The bile duct joins the pancreatic duct.

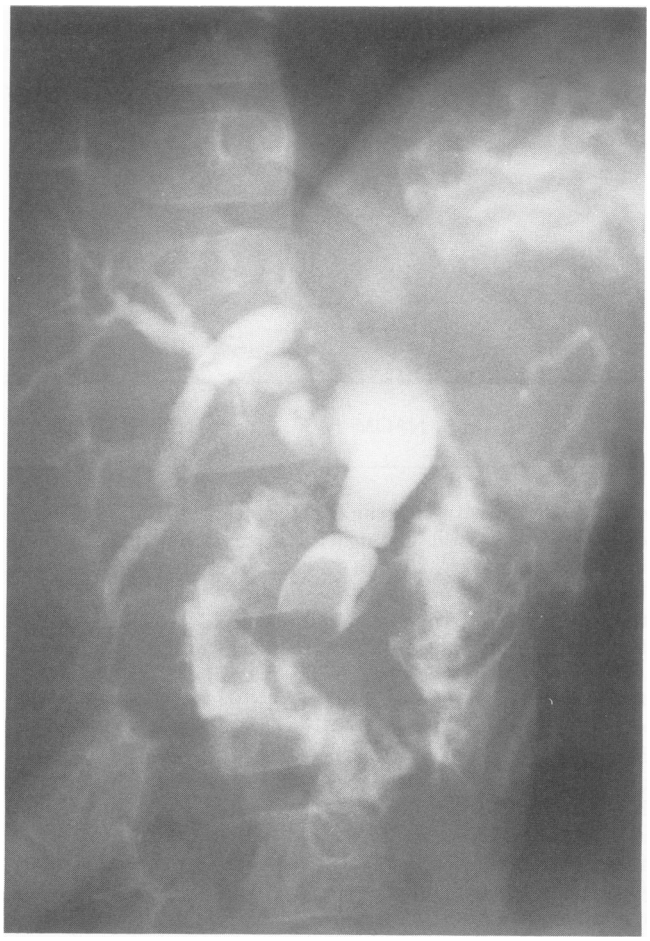

FIG. 2. P-C type of abnormal choledochopancreaticoductal junction. The pancreatic duct joins the bile duct.

Informed consent was obtained from the parents before performing intraoperative biliary manometry.

Results

Types of Abnormal Choledochopancreaticoductal Junction in Relation to Forms of Choledochal Dilatation

Twenty-six patients with congenital choledochal dilatation could be analyzed by preoperative ERCP or by operative cholangiography whether they had an abnormal choledochopancreaticoductal junction or not (Table 2). An abnormal choledochopancreaticoductal junction was noted in 25 of the 26 patients (96.2%) with congenital choledochal dilatation. The remaining patient (3.8%), with a fusiform choledochal dilatation, had no abnormal choledochopancreaticoductal junction.

Of the 12 patients with cystic dilatation, 10 had the C-P type, 1 had the P-C type, and 1 had the miscellaneous

TABLE 2. *Forms of Choledochal Dilatation and Abnormal Choledochopancreaticoductal Junction*

Form of Choledochal Dilatation	No. of Patients	Abnormal Choledochopancreaticoductal Junction			
		C-P type	P-C type	Miscellaneous	Absent
Cystic	12	10	1	1	—
Fusiform	13	2	9	1	1
Multicystic	1	—	—	1	—
Total	26	12	10	3	1

TABLE 3. *Forms of Choledochal Dilatation and Amylase Levels in Cyst and Gallbladder*

Form of Choledochal Dilatation	No. of Patients	Amylase Levels	
		Cyst	Gallbladder
Cystic	22	59,969 ± 14,311	80,065 ± 18,724
Fusiform	13	42,160 ± 9091*	91,284 ± 24,238*
Multicystic	2	NE	200,010.0

Mean ± SE.
* $0.05 < p < 0.1$
NE, not examined.

type of junction. Of the 13 patients with fusiform dilatation, 9 had the P-C type, 2 had the C-P type, and 1 had the miscellaneous type of junction, and 1 had a normal junction. One patient had a multicystic type of dilatation and the miscellaneous type of abnormal choledochopancreaticoductal junction.

Amylase Levels in Choledochal Cyst and Gallbladder

In 22 patients with cystic dilatation the mean amylase levels in the choledochal cyst and the gallbladder were 59,969 ± 14,311 Somogyi Units and 80,065 ± 18,724 Somogyi Units, respectively (Table 3). In 13 patients with fusiform dilatation, the mean amylase level in the gallbladder (91,284 ± 24,238 Somogyi Units) was higher ($0.05 < p < 0.1$) than that in the choledochal cyst (42,160 ± 9,091 Somogyi Units). In two patients with multicystic type dilatation, the mean amylase level in the gallbladder was 200,010 Somogyi Units, and the amylase level in the choledochal cysts could not be examined.

Pressure Measurements in the Biliary Tract Before and After Gastrin Stimulation

Before gastrin stimulation the mean pressures in the duodenum and sphincter of Oddi were 9.4 ± 1.2 cm H_2O and 32.0 ± 3.7 cm H_2O, respectively (Table 4). The difference between the sphincter of Oddi pressure and the duodenal pressure was 22.8 ± 3.1 cm H_2O. The length of the high-pressure zone in the sphincter of Oddi was 1.6 ± 0.2 cm. The choledochopancreaticoductal junction pressure was 12.8 ± 1.6 cm H_2O.

The mean sphincter of Oddi pressure in the cystic type and in the fusiform type after gastrin stimulation was 45.4 ± 4.1 cm H_2O and 41.0 ± 3.3 cm H_2O, respectively. These values were significantly higher ($p < 0.05$, $0.05 < p < 0.1$) than before gastrin stimulation. However, there was no significant difference between these two types. The mean differences between sphincter of Oddi pressure and duodenal pressure in the cystic type and in the fusiform type after gastrin stimulation were 37.0 ± 4.2 cm H_2O and 34.0 ± 3.7 cm H_2O, respectively. These values were significantly higher ($p < 0.05$) than before gastrin stimulation. However, there was no significant difference between these two types. The mean lengths in the cystic type and in the fusiform type after gastrin stimulation were 1.6 ± 0.3 cm and 1.7 ± 0.2 cm, respectively. The choledochopancreaticoductal junction pressures in the cystic type and in the fusiform type after gastrin stimulation were 13.0 ± 1.1 cm H_2O and 13.5 ± 0.8 cm H_2O, respectively. Thus, tetragastrin stimulation caused no significant change in choledochopancreaticoductal junction pressures before and after gastrin stimulation.

Discussion

As the cause of choledochal cyst, Babbit[2] proposed an abnormal relationship between the common bile duct and

TABLE 4. *Pressure Measurements of Biliary Tract in Cystic and Fusiform Types Before and After Tetragastrin Stimulation*

	Duodenal Pressure (cm H_2O)	Sphincter of Oddi Pressure (cm H_2O)	Difference Between Sphincter of Oddi Pressure and Duodenal Pressure (cm H_2O)	Length of High-Pressure Zone (cm)	Choledochopancreaticoductal Junction Pressure (cm H_2O)
Before stimulation (n = 11)	9.4 ± 1.2	32.0 ± 3.7*†	22.8 ± 3.1‡	1.6 ± 0.2	12.8 ± 1.6
Cystic type after gastrin stimulation (n = 7)	8.4 ± 1.3	45.4 ± 4.1*	37.0 ± 4.2‡	1.6 ± 0.3	13.0 ± 1.1
Fusiform type after gastrin stimulation (n = 4)	7.0 ± 1.1	41.0 ± 3.3†	34.0 ± 3.7‡	1.7 ± 0.2	13.5 ± 0.8

Mean ± SE.
* $p < 0.05$.
† $0.05 < p < 0.1$.
‡ $p < 0.05$.

the pancreatic duct. Miyano et al[3] and Kimura et al[4] reported that all of the patients with choledochal cyst, except those with choledochocele, had an abnormal choledochopancreaticoductal junction. Komi et al[5] also reported the frequency of abnormal anomalous ductal union in patients with choledochal cyst to be 39%. In our series, 25 of the 26 patients (96.2%) with congenital choedochal dilatation had an abnormal choledochopancreaticoductal junction. Therefore, there is no denying the fact that choledochal dilatation has a close association with abnormal choledochopancreaticoductal junction, whether direct or indirect.

Types of abnormal choledochopancreaticoductal junction were analyzed in relation to the forms of choledochal dilatation. In our series, the C-P type was often found in the cystic type of choledochal dilatation, and the P-C type was often found in the fusiform type of choledochal dilatation. These results are consistent with those previously reported.[6,7] Therefore, the types of abnormal choledochopancreaticoductal junction seem to correlate well with the degree of choledochal dilatation.

The present study showed that the amylase levels in the choledochal cyst and the gallbladder were elevated regardless of the form of choledochal dilatation. These results indicate that a free reflux of pancreatic juice into the biliary system occurs in patients with congenital choledochal dilatation. In patients with the fusiform type of choledochal dilatation, the amylase level in the gallbladder was significantly higher than in the choledochal cyst. This result is natural, because the gallbladder is said to concentrate bile to between 10% and 20% of its original volume.[8]

Kimura et al.[4] stressed that regurgitation of pancreatic juice was an important factor in gallbladder carcinoma, especially in patients with anomalous pancreaticobiliary ductal union. In our series, we actually had a patient with adenocarcinoma, a 12-year-old girl with anomalous pancreaticobiliary ductal union. From these results, it seems reasonable to assume that the longstanding inflammation of the biliary tract caused by the reflux of pancreatic juice might be one of the factors in carcinogenesis in the biliary tract, especially in the gallbladder.

Intraoperative manometry showed a high-pressure zone in the area of the sphincter of Oddi in patients with congenital choledochal dilatation. However, no high-pressure zone was found in the area of the common channel or the choledochopancreaticoductal junction. This indicates that the influence of the sphincter of Oddi pressure does not extend to the area of the abnormal choledochopancreaticoductal junction. Gastrin increased the sphincter of Oddi pressure in patients with congenital choledochal dilatation. No increase of pressure was found in the area of the common channel or the choledochopancreaticoductal junction, however, even after the administration of gastrin. This indicates that the change in sphincter pressure caused by gastrin administration does not extend to the area of abnormal choledochopancreaticoductal junction.

These results suggest that a free reflux of pancreatic juice into the biliary system can occur, and this free reflux may be due to the lack of sphincter function at the junction of the common bile and pancreatic ducts.

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